

# Hemi-Fontan Procedure

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The evolution of the Fontan procedure for single ventricle cardiac malformations has included the development of several surgical modifications that appear to have decreased overall morbidity and mortality. The inclusion of a second-stage intervention to create a cavopulmonary source of blood flow to decrease the volume load on the single ventricle and decrease the effect of ventricular hypertrophy and diastolic compliance abnormalities on late function of the Fontan operation resulted from the development of both the bidirectional Glenn shunt and hemi-Fontan modifications.<sup>1-5</sup> In the early experience with the hemi-Fontan procedure, several surgical modifications have been described. The technique used at the Children's Hospital of Philadelphia was developed by Drs Norwood and Jacobs in the late 1980s and continues to be used at our institution.<sup>3</sup> The advantage of the hemi-Fontan procedure is the significant augmentation of the branch pulmonary arteries, which eliminates any area of narrowing related to the previous placement of aortopulmonary shunts. In addition, the technique maintains a cavoatrial connection with a homograft dam, which can be reopened at the time of completion of lateral tunnel Fontan operation. The technique was developed at a time when the lateral tunnel Fontan technique was the predominant technique used for completion of Fontan's operation and had the advantage of more streamlined flow through the lateral aspect of the atrium to the pulmonary arteries, which improved overall hydrodynamic efficiency of the Fontan operation.

In addition to the beneficial effect on pulmonary artery augmentation, the hemi-Fontan procedure facilitates the completion of the lateral tunnel Fontan procedure and also creates an anteroposterior offset of the superior vena caval and pulmonary arterial flows, which may have some hemodynamic benefit.<sup>6</sup> In addition, the fenestrated lateral tunnel Fontan procedure in our institution has been associated with a decreased incidence and duration of pleural effusions.<sup>7</sup>

The hemi-Fontan operation has been associated with low operative and intermediate-term mortality. Initial reports of the hemi-Fontan operation at the Children's Hospital of Philadelphia had initial mortality rates as high as 8%; however, over the course of the last 10 years, there has been progressive decrease in operative mortality such that

currently the mortality for elective hemi-Fontan operation is 1% or less. Including patients with multiple risk factors, the mortality has been 2.6% in 154 patients from 1995 through 2013. This mortality level is comparable to the mortality for the bidirectional Glenn shunt in our own institution. Currently both the hemi-Fontan and bidirectional Glenn shunt are used at the Children's Hospital of Philadelphia, primarily based on surgeon preference and individual patient anatomy. The bidirectional Glenn shunt is utilized by some surgeons preferentially; however, others use the hemi-Fontan in patients who require extensive pulmonary arterioplasty for pulmonary artery distortion, which has become increasingly common with the RV to PA shunt modification of the Stage I Norwood operation.

The selection of the second-stage shunting procedure determines the technique utilized for the completion Fontan operation. When a bidirectional Glenn shunt is utilized then an extracardiac completion Fontan is performed, as there is no point in reconnecting the superior vena cava to the right atrium after a bidirectional Glenn shunt. Patients who have the hemi-Fontan modification have an anatomy that is extremely suitable for completion by the lateral tunnel Fontan operation, which is preferentially used in these patients. There is little reason to perform an extracardiac Fontan completion after hemi-Fontan procedures as the dam created between the superior vena cava and right atrium is ideally suited to be removed to create a large opening between the superior vena cava and right atrium adequate for total cavopulmonary flow.

In the Children's Hospital of Philadelphia experience the mortality at Fontan completion is similarly low between the lateral tunnel and extracardiac techniques with a hospital mortality of around 1% and 1% incidence of Fontan take down or transplantation. In an experience of more than 770 consecutive Fontan patients in our institution with contemporaneous usage of both the lateral tunnel and extracardiac Fontan procedures with routine fenestration, the duration of pleural effusions was slightly higher in the extracardiac Fontan group, primarily owing to a less effective fenestration.<sup>7</sup> The lateral tunnel Fontan produced a very effective and consistent fenestration, which decreased the duration of pleural effusions and subsequent hospital stay.

A major disadvantage of the hemi-Fontan procedure is the fact that the sinus node artery is often divided at the time of creation of the hemi-Fontan baffle. Although this would seem to be associated with arrhythmias, studies at the Children's Hospital of Philadelphia and elsewhere have shown that although the incidence of sinus rhythm may be

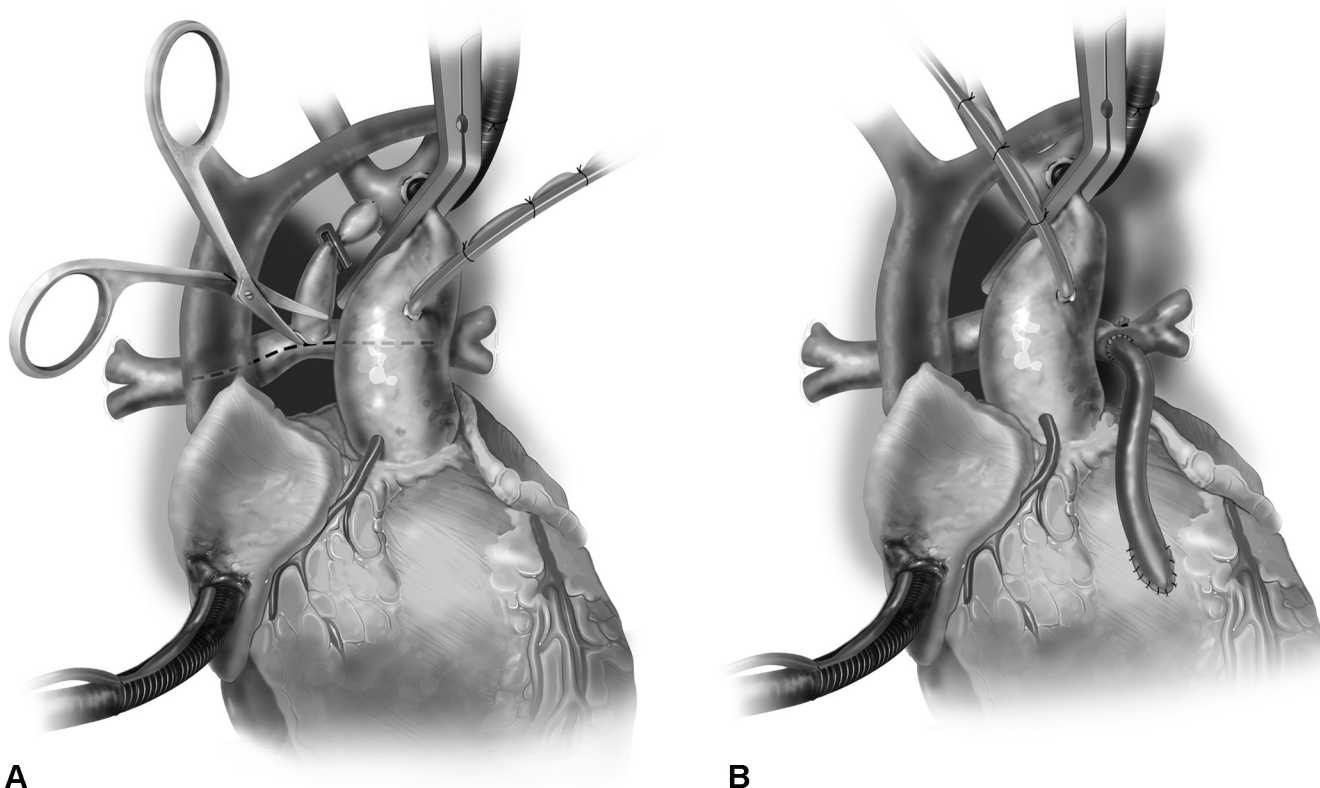
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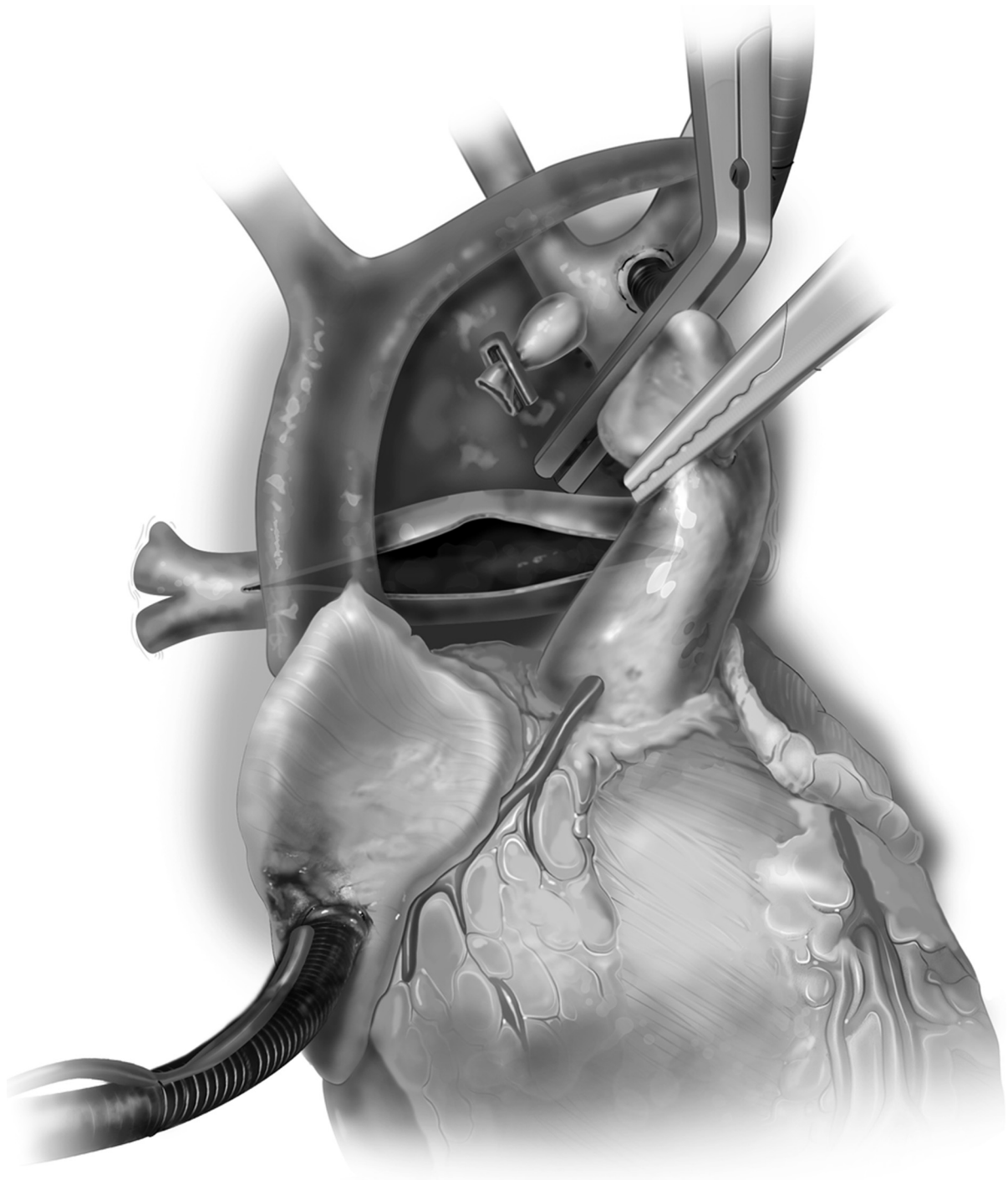
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slightly decreased in the immediate perioperative period, by discharge the vast majority of patients have returned to a normal sinus rhythm, which is maintained.<sup>8</sup> Comparisons of the incidence of sinus rhythm late after the Fontan operation by either lateral tunnel or extracardiac technique in our institution have shown no demonstrable difference in late sinus node dysfunction between the 2 procedures.<sup>9</sup> Thus it appears that although there is a potential disadvantage of interference with sinus node blood supply, there are other factors that may have more importance for the development of late sinus node dysfunction or junctional rhythm. It is possible that mobilization of the superior vena cava and interference with the neural supply to the area of the sinus node may be more important than sinus node blood supply, which can be augmented directly from the right atrial chamber. This subject needs to be investigated more fully (Figs. 1-12).

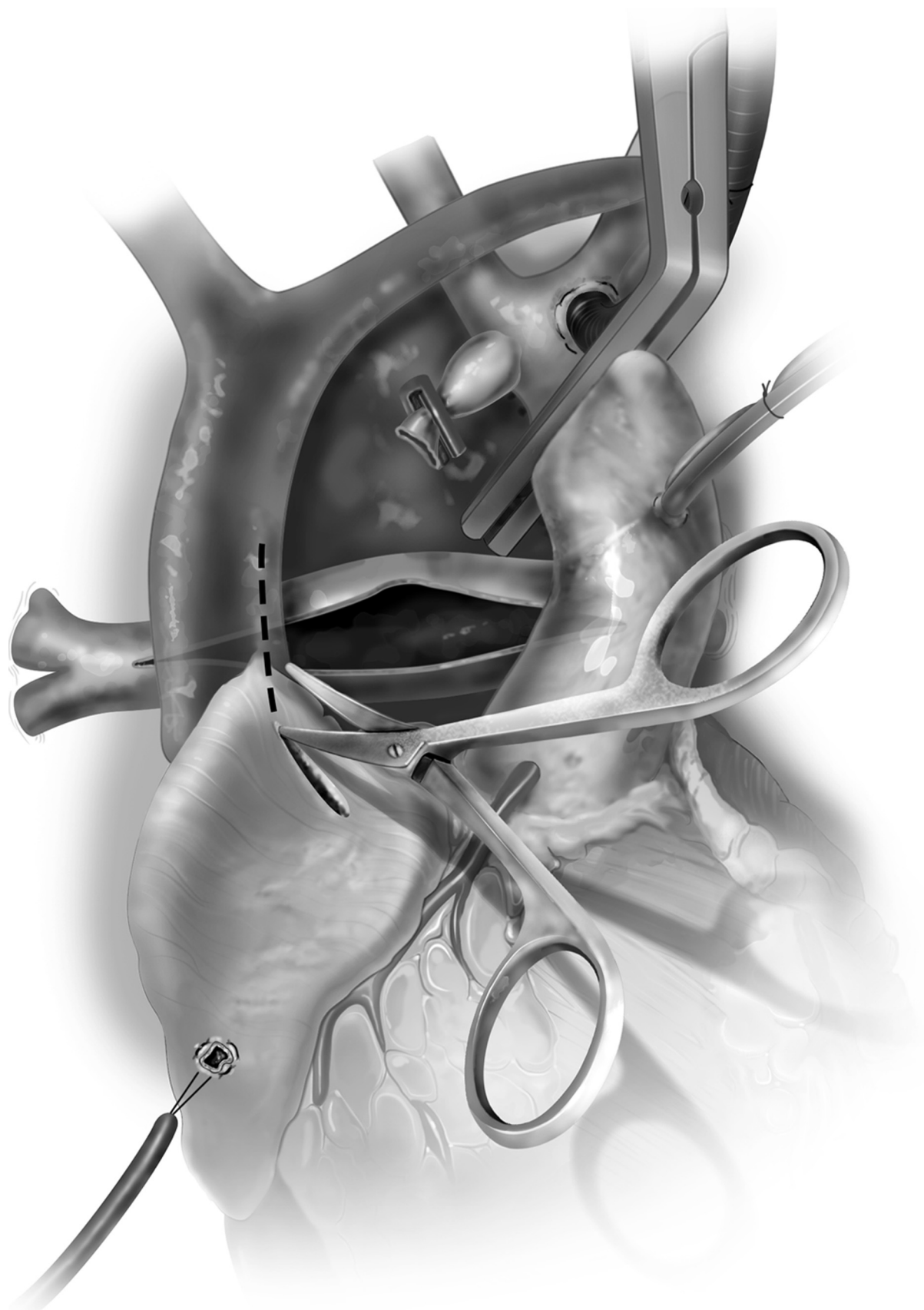
There has been some controversy about the hemodynamic benefits of the hemi-Fontan vs the bidirectional Glenn shunt. Computational fluid dynamic studies have suggested that the anteroposterior offset of the superior vena caval and pulmonary arterial flows in the hemi-Fontan procedure decreases energy loss and improves distribution of blood flow between the pulmonary arteries.<sup>6</sup> Other studies have suggested that the bidirectional Glenn shunt may have more hemodynamic advantage. However, the relative contributions of fluid dynamics to late function of the Fontan operation have not been confirmed. Other studies comparing the lateral tunnel and extracardiac Fontan procedure have suggested that the use of the hemi-Fontan and lateral tunnel Fontan operation have similar instances of sinus node dysfunction and overall mortality; however, the lateral tunnel Fontan after the hemi-Fontan procedure uses less hospital resources primarily because of decreased duration of pleural effusions.<sup>10</sup>



**Figure 1** (A) The median sternotomy incision used for the initial stage I reconstruction is reopened and adhesions are taken down over the atrium, ventricle, and the neo-aorta. The right modified Blalock-Taussig shunt is mobilized along with the branch pulmonary arteries behind the neo-aorta. If a modified Blalock-Taussig shunt was utilized for the initial procedure, the patient is placed on cardiopulmonary bypass, generally with a single aortic and venous cannula and the use of deep hypothermic circulatory arrest. After cooling to 18°C, circulatory arrest is established, the neo-aorta clamped and cardioplegia administered into the neo-aortic root. The modified Blalock-Taussig shunt is ligated and clipped as proximally as possible on initiation of bypass to prevent pulmonary flow during cooling. After circulatory arrest is established and cardioplegia administered, the shunt is divided from the entrance into the right pulmonary artery and a segment of shunt material removed. As shown in 1A, an incision is then made widely to the hilum of the lung on the left at the take off of the left upper lobe branch, behind the neo-aorta and then on the right pulmonary artery to the origin of the right lower lobe. (B) In patients who have had a previous right ventricle to pulmonary artery shunt placed at the initial Norwood operation, again, circulatory arrest is utilized and after cardioplegia is administered the right ventricle to pulmonary arterial shunt is ligated as close as possible to the origin from the right ventricle and divided at the pulmonary bifurcation distally. Again, an incision is made from hilum to hilum on the pulmonary artery extending to the left upper lobe branch and to the right lower lobe branch of the pulmonary vessel.

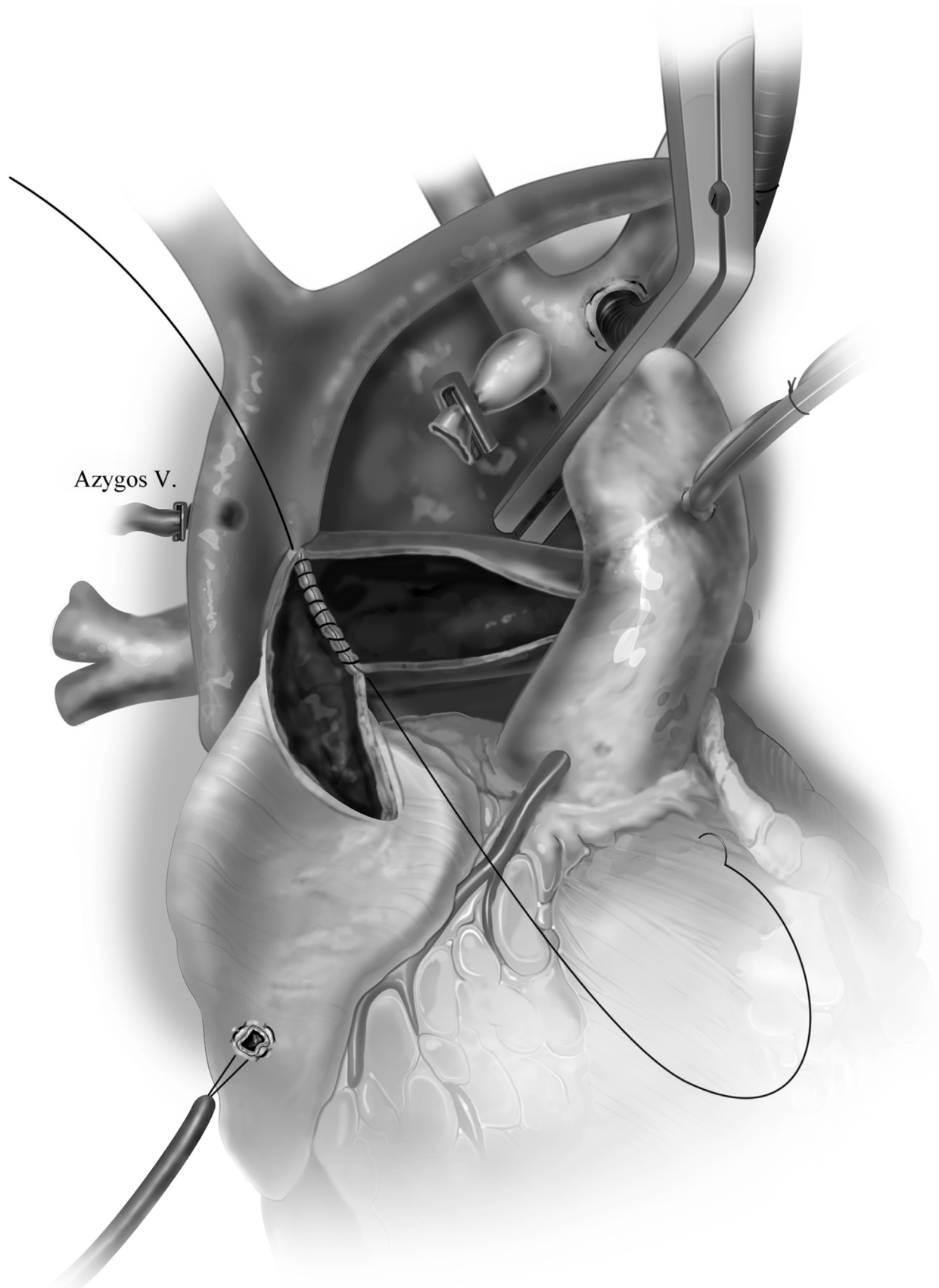


**Figure 2** After opening of the pulmonary arteries extensively, any areas of narrowing are opened such that the pulmonary bifurcation is completely unobstructed.

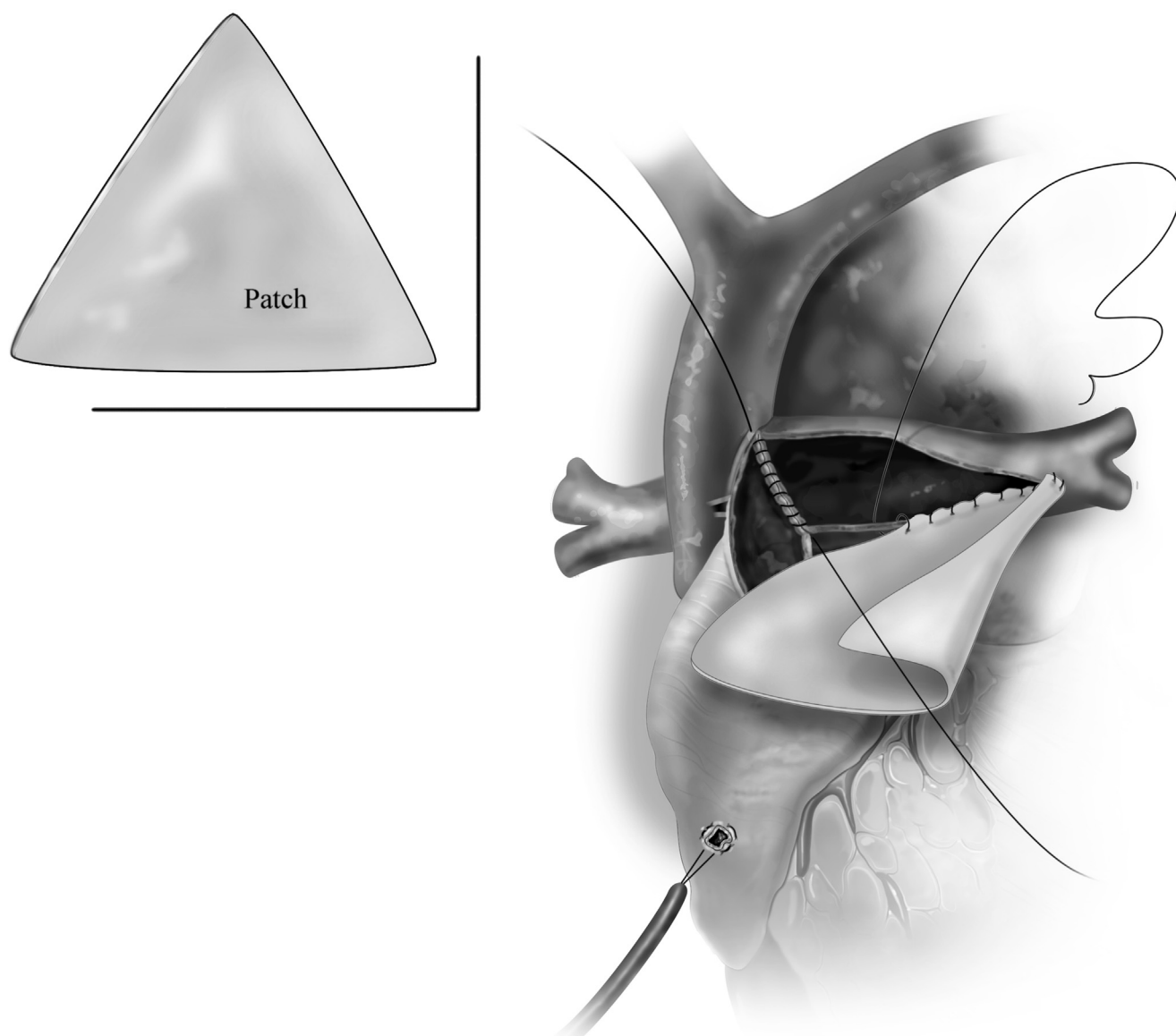


**Figure 3** An incision is then made in the anterior aspect of the right atrial appendage and the incision then carried medially across the cavoatrial junction for approximately 1 cm onto the superior vena cava using scissors. In most cases, this incision divides the sinus node artery if it is located anteriorly. In our experience this has not resulted in any significant incidence of sinus node dysfunction and late evaluation of sinus node function has been similar between the hemi-Fontan operation and the bidirectional Glenn shunt, which does not generally interfere with the sinus node artery.

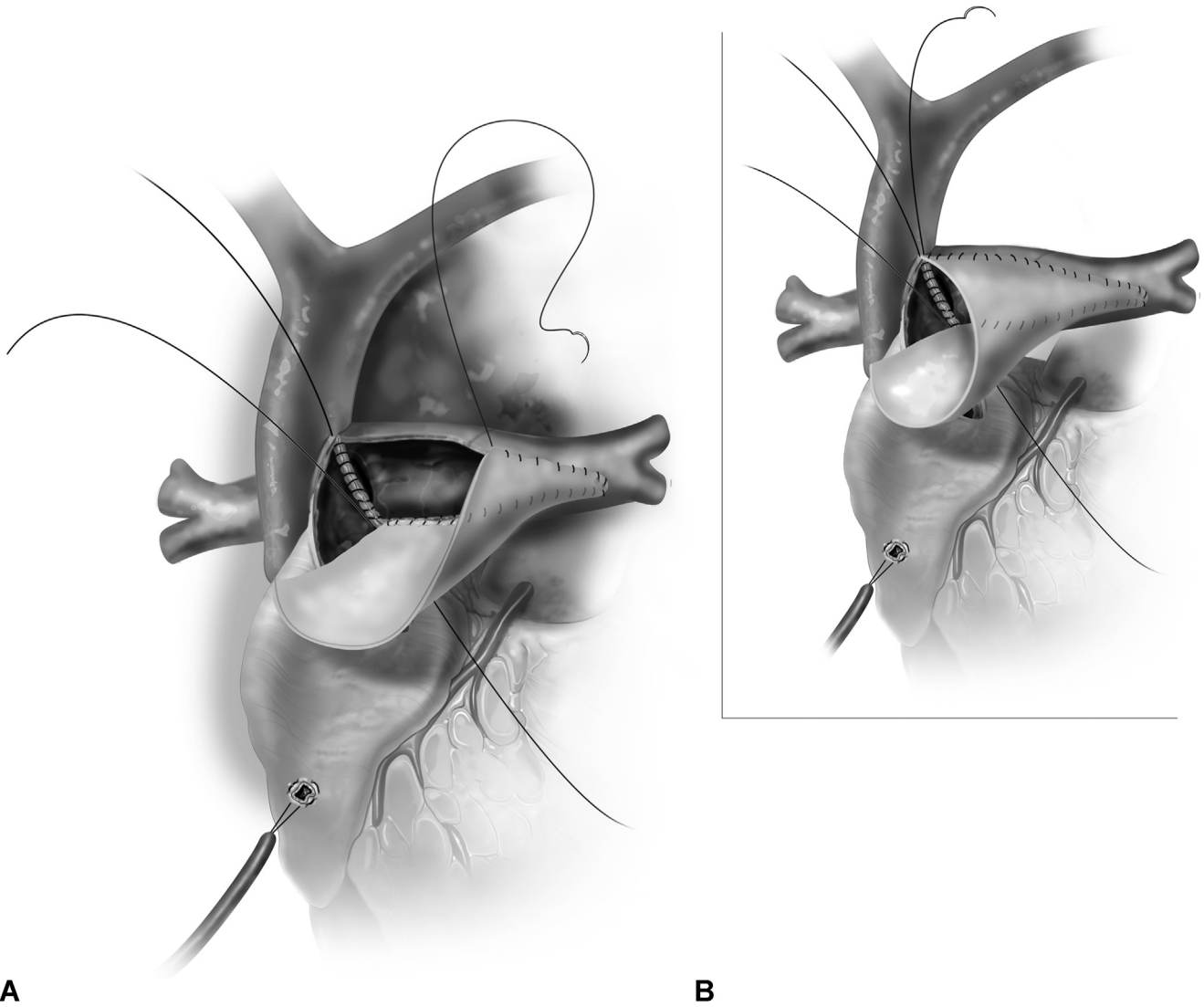




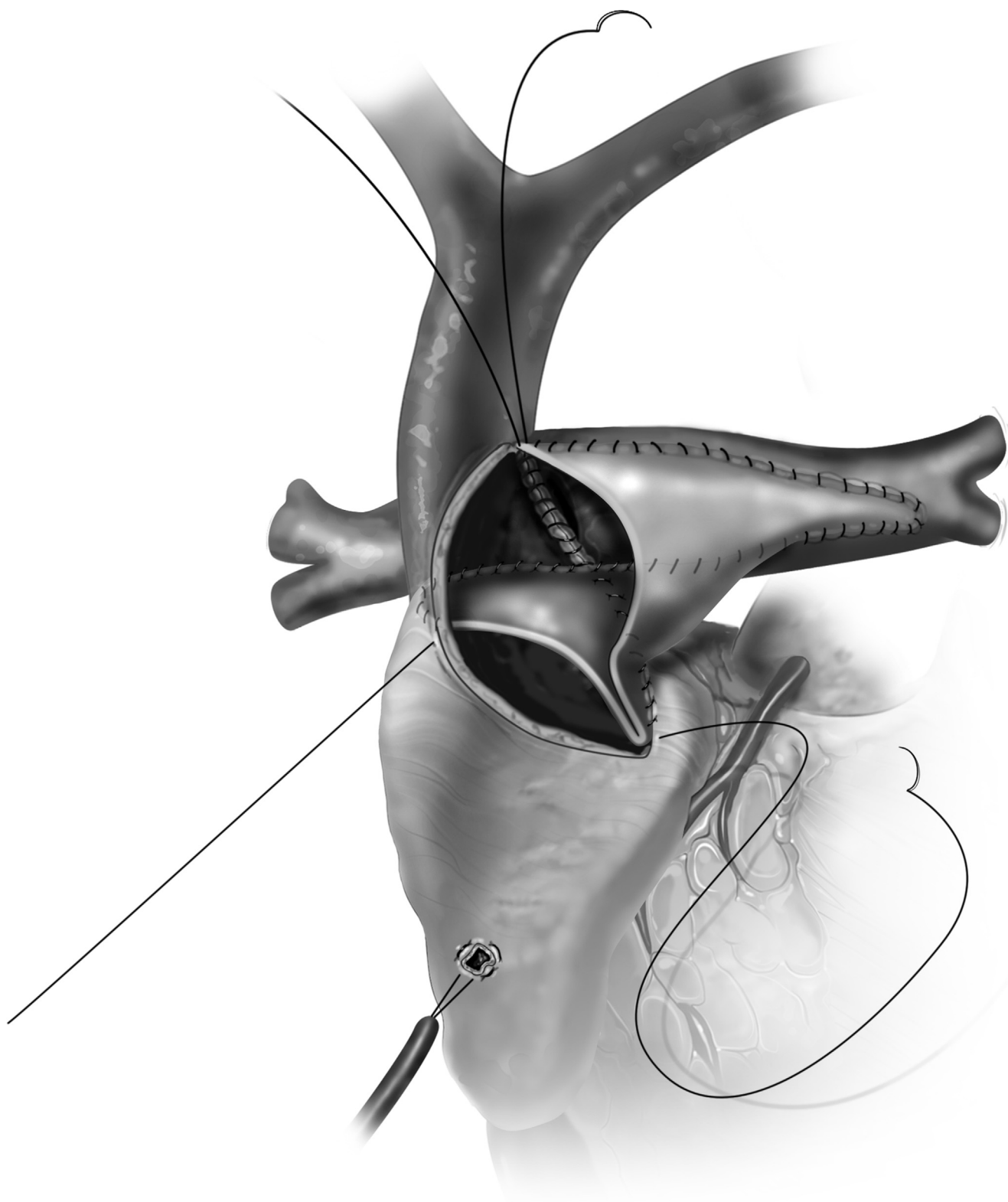
**Figure 4** The posterior aspect of the superior vena cava and right atrial opening is then sutured to the anterior aspect of the pulmonary artery opening using a running fine polypropylene suture. In this fashion, the flow from the superior vena cava will pass over this suture line into the central pulmonary arteries and then posteriorly into the right pulmonary artery. This creates an offset between the superior vena cava and pulmonary artery, which has been associated with better flow distribution and lack of energy loss compared with a direct end-to-side connection as in the bidirectional Glenn shunt. V = vein.



**Figure 5** A patch of pulmonary homograft material is cut in a wide triangular shape with as broad a base as possible. Generally a portion of pulmonary homograft material called a hemipatch is utilized for this. The patch is sutured at the leftward extent of the pulmonary arterial incision behind the neo-aorta and brought over to the anastomosis of the superior vena cava and right pulmonary artery as shown in this figure.

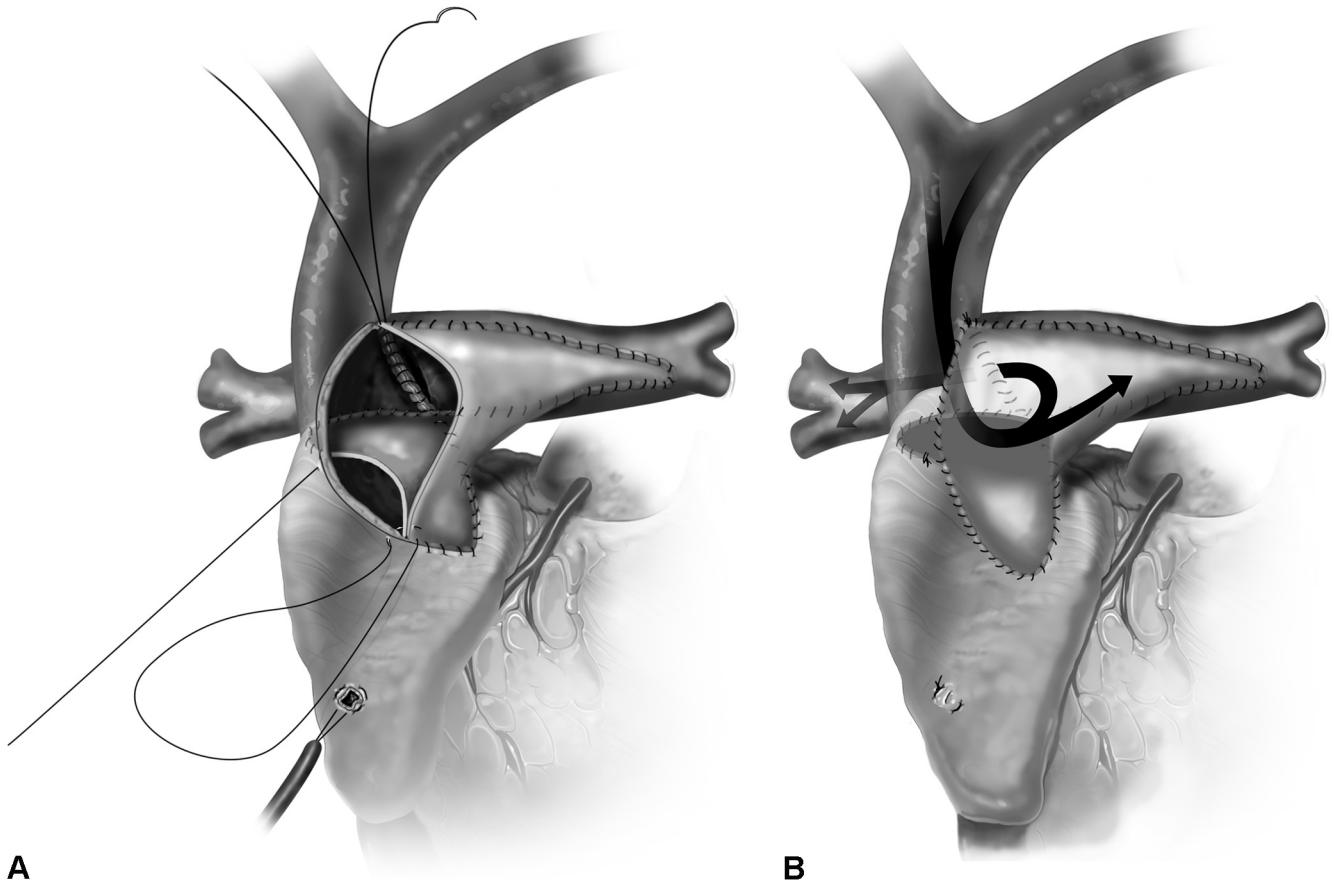


**Figure 6** (A) As the patch is sewn to the superior aspect of the widely opened pulmonary artery, the suture line is carried to the previous suture line joining the superior vena cava and right atrium to the right pulmonary artery and the sutures are tied at this point. (B) With this wide patching of the pulmonary arteries, extensive enlargement of the pulmonary vessels is created and any areas of minor obstruction are dealt with.

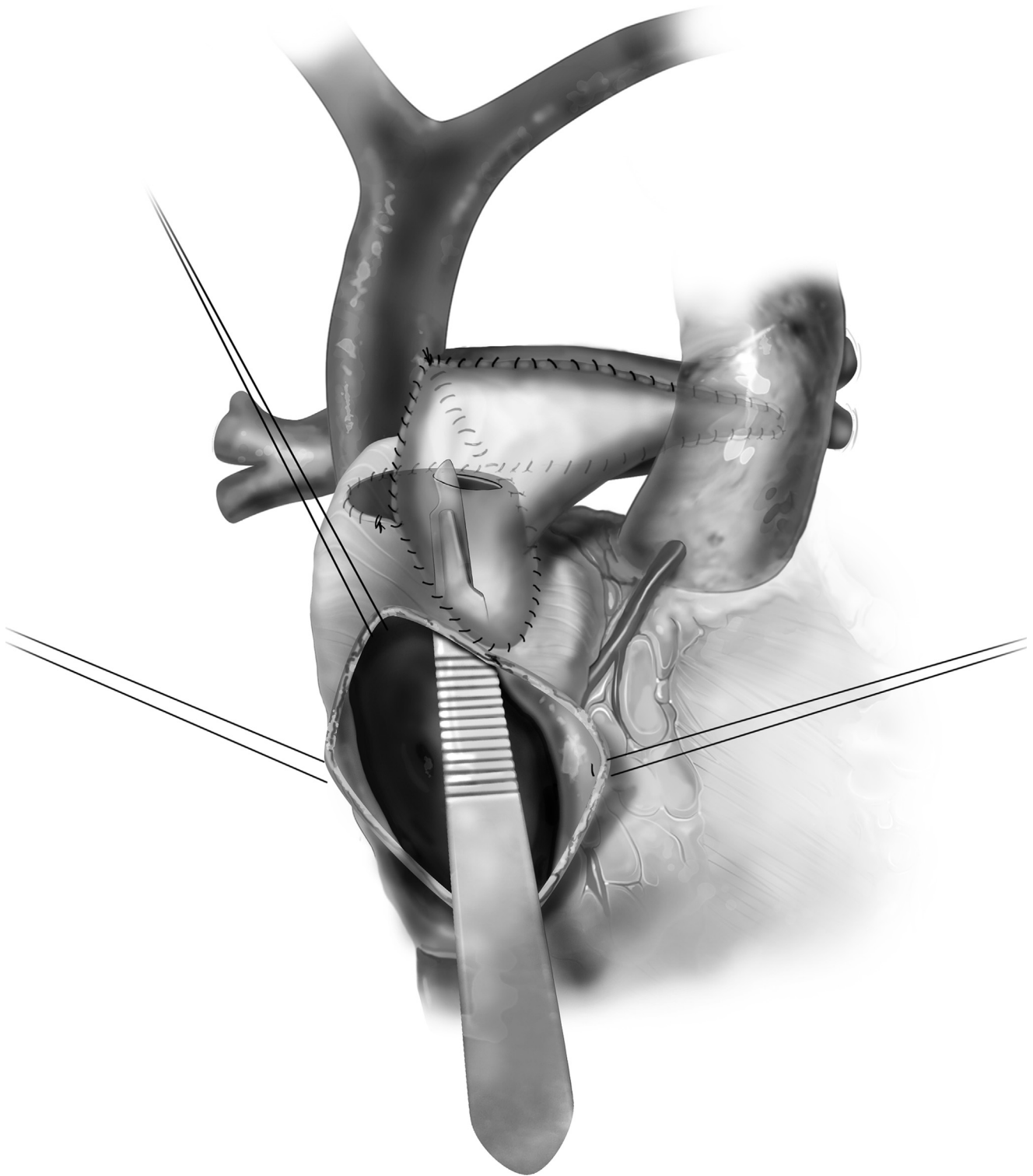


**Figure 7** The suture line inferiorly along the pulmonary artery is then brought down and carried into the junction between the superior vena cava and right atrium as shown in this figure. In this fashion a “dam” of pulmonary homograft material is used to block off the superior vena cava from the right atrium. In this figure, as shown, the wide patch of pulmonary homograft material is folded down to create this dam between the superior vena cava and right atrium, generally using a running 5-0 polypropylene suture.

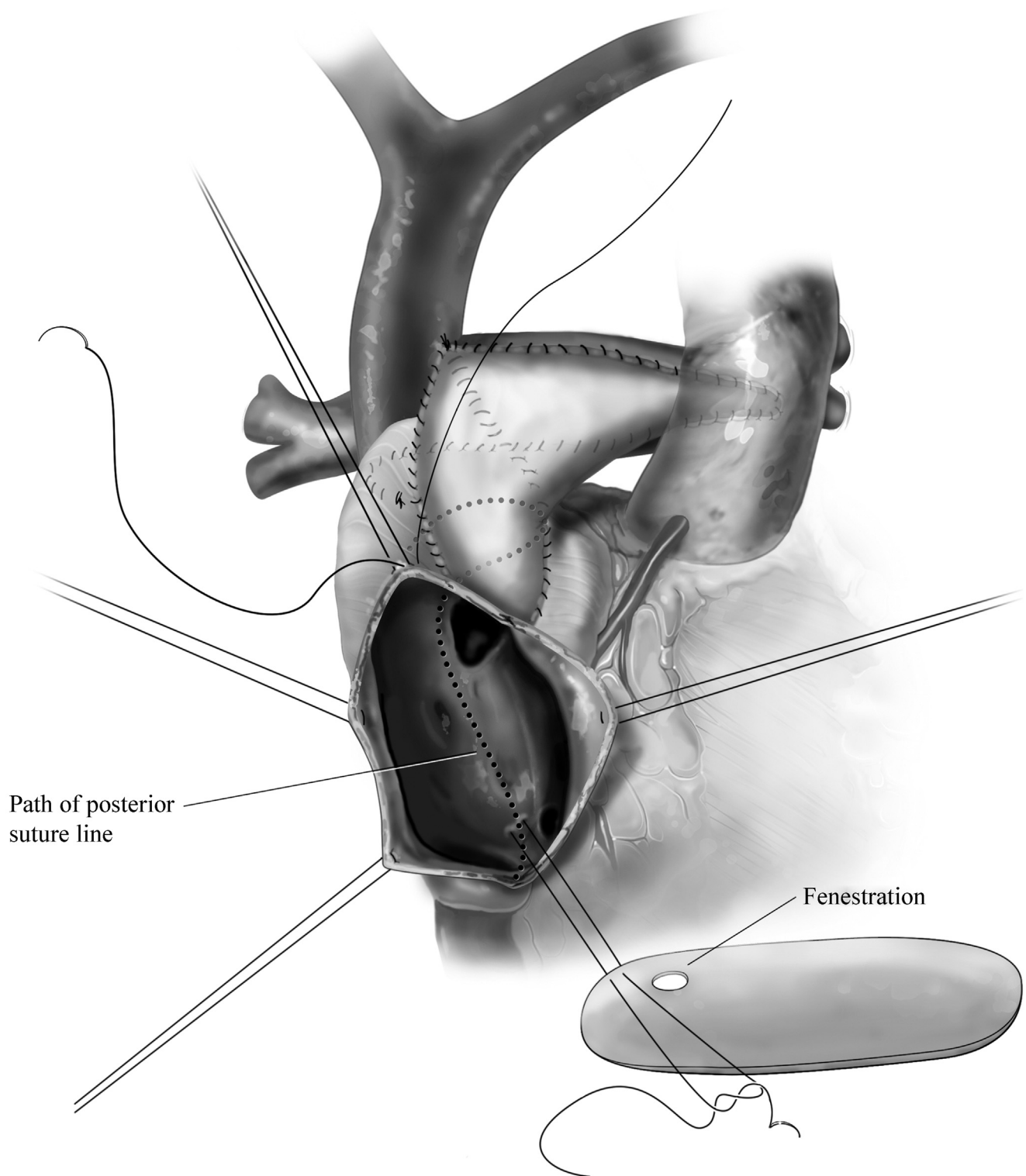




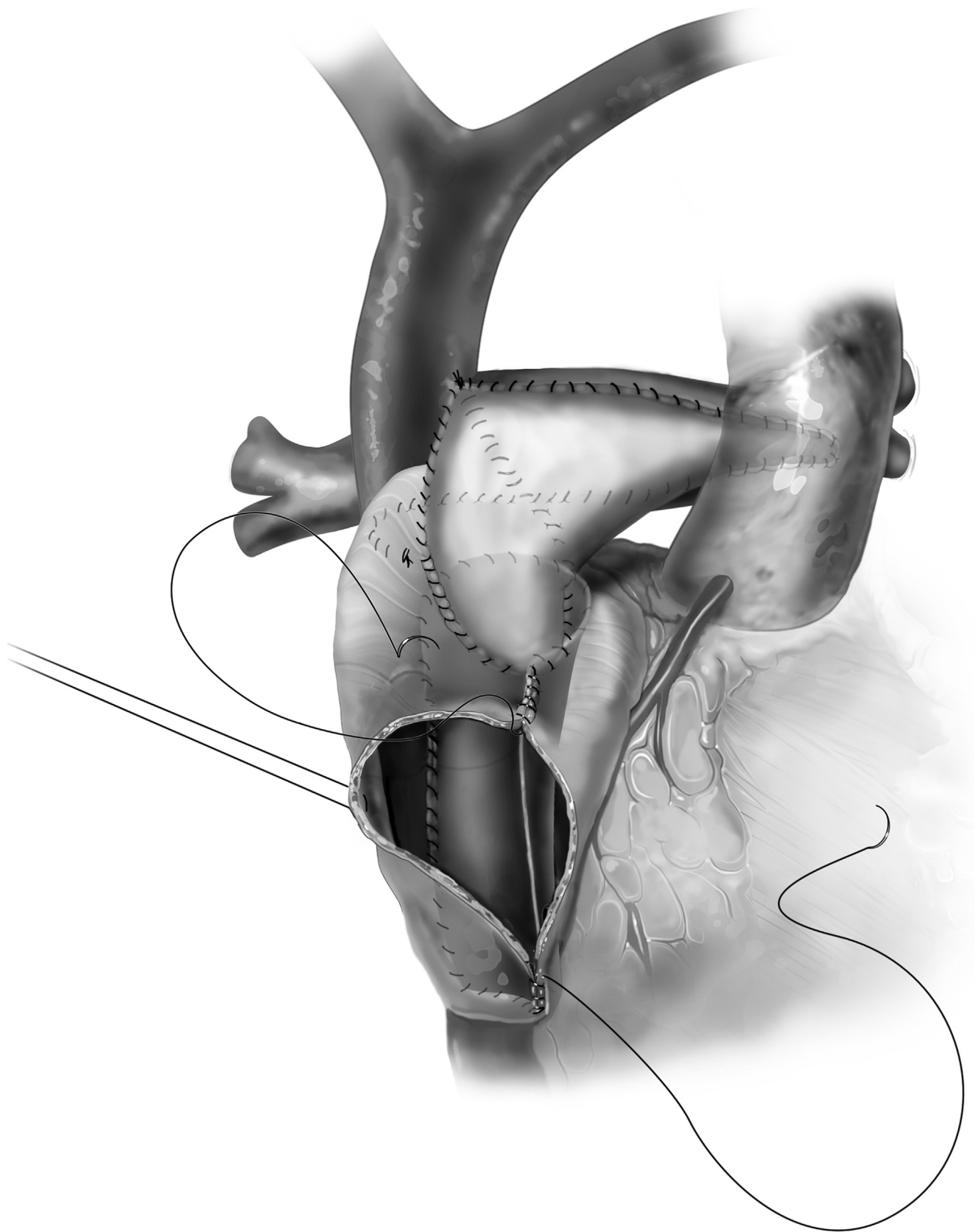
**Figure 8** (A) As the suture line is continued and the patch is folded down, the superior vena caval flow is completely blocked from the right atrium leaving a floor of homograft material, which can then be excised for the completion of Fontan operation. (B) At the completion of the anastomosis with the remaining suture line onto the superior vena cava completed, flow from the superior vena cava comes down, is blocked by the dam between the superior vena cava and right atrium, and then flows into the main pulmonary artery and posteriorly into the right pulmonary artery as shown in this diagram.



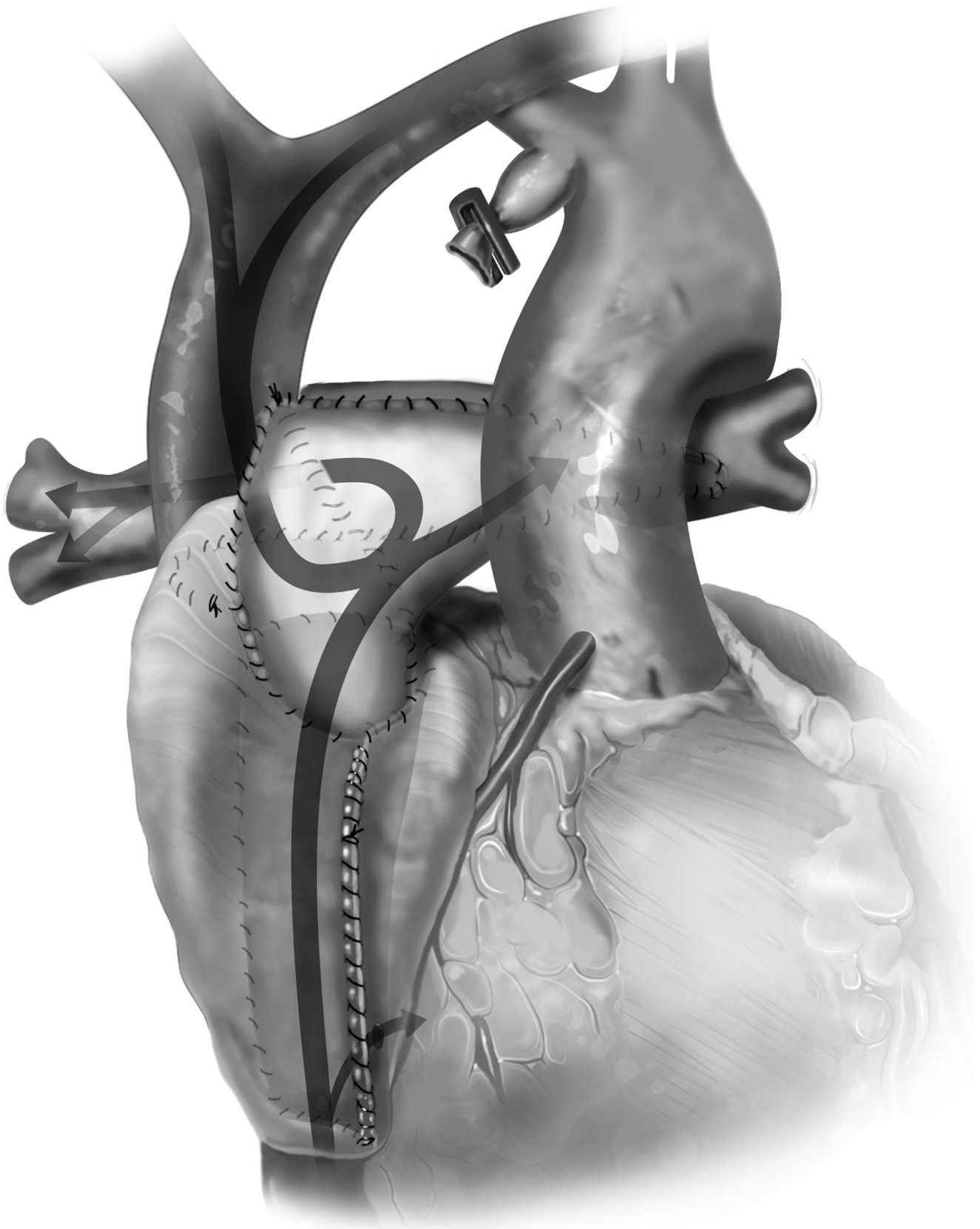
**Figure 9** Fontan operation: At the time of the Fontan operation, again, deep hypothermia and circulatory arrest is generally utilized in our center. A longitudinal incision is made in the right atrium from the medial aspect of the inferior vena cava superiorly to the edge of the homograft dam between the superior vena cava and right atrium. Working through the right atrium, the dam of homograft material is excised as shown in this figure, reopening the connection between the superior vena cava and right atrium for completion of the Fontan connection. Because of the initial incision across the cavoatrial junction, the dam of homograft material is larger in diameter and area than the inferior vena cava, thus preventing any area of stenosis or flow restriction with the lateral tunnel connection into the pulmonary arteries.



**Figure 10** After excision of the dam of homograft material, generally a 10-mm Impra® or Gortex® graft is incised longitudinally and trimmed and a 4-mm fenestration created in the graft material. This baffle of polytetrafluoroethylene material is then sutured into the atrium, away from the atrial septal defect and then inferiorly around the eustachian valve up to the opening in the right atrium. Superiorly the baffle is then sutured around the previous suture line that created the dam of tissue between the superior vena cava and right atrium. Because of this previous suture line, suturing in this area is quite straightforward, and there is little concern of leakage around pectinate muscles in the atrium, as this is an area of previous scar tissue. The suture line is carried inferiorly around the previous suture line and then anteriorly along the suture line to the superior aspect of the right atrial incision.



**Figure 11** Once the atrial baffle is completed to the suture line in the atrium the anterior suture line of the baffle to the atrium is used with the atrial closure to complete the lateral tunnel Fontan repair.



**Figure 12** At the completion of the lateral tunnel Fontan procedure, flow from the inferior vena cava partially crosses the fenestration to the single ventricle and the remaining flow follows the lateral aspect of the right atrium to the pulmonary arteries. Superior venal caval flow is baffled to the main pulmonary arteries and posteriorly to the right pulmonary artery creating anteroposterior offset to the cavopulmonary connection.



## Comments

The hemi-Fontan procedure as described in this review as performed at the Children's Hospital of Philadelphia has become a standardized procedure at our institution. It is preferentially utilized when pulmonary artery augmentation is desirable at the second-stage reconstruction and can be performed at the same age as a bidirectional Glenn shunt. The creation of a homograft dam between the superior vena cava and right atrium allows for easy completion of lateral tunnel Fontan operation and the opening between the cava and atrium is larger than the original superior vena caval opening, which prevents obstruction of flow from the inferior vena cava to the pulmonary arteries. With completion lateral tunnel Fontan connection, growth of the lateral tunnel can occur owing to the native right atrial wall incorporated into the lateral tunnel connection. Thus, the lateral tunnel completion operation can be performed at essentially any age and body weight.

We have elected to perform our second- and third-stage procedures at the Children's Hospital of Philadelphia under a short period of deep hypothermic circulatory arrest. The simplicity of these operations and their reproducibility have resulted in a very consistently low and safe period of deep hypothermic circulatory arrest with no evidence of increased neurologic abnormalities. The hemi-Fontan procedure can generally be performed with a circulatory arrest time of less than 25–30 minutes and the lateral tunnel completion operations can be performed with circulatory arrest times of less than 20 minutes in virtually all situations.

Continued longitudinal follow-up of the Fontan patients with the various techniques of second- and third-stage reconstruction will have to be performed to see if there are any late differences in overall survival or

complications between the various techniques. At the present time both hemi-Fontan and lateral tunnel Fontan procedures and bidirectional Glenn shunt with extracardiac Fontan procedures are performed commonly in our institution based on individual patient anatomy and surgeon preference.

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